Surgical outcomes of 380 patients with double outlet right ventricle who underwent biventricular repair

Shoujun Li, MD, Kai Ma, MD, PhD, Shengshou Hu, MD, Zhongdong Hua, MD, Keming Yang, MD, Jun Yan, MD, and Qiuming Chen, MD, PhD

Objectives: The study objective was to report the outcomes of biventricular repair in patients with double outlet right ventricle.

Methods: Patients with double outlet right ventricle who underwent biventricular repair at Fuwai Hospital from January 2005 to December 2012 were included. Patients were excluded if double outlet right ventricle was combined with atrioventricular septal defect, heterotaxy syndrome, atrioventricular discordance, or univentricular physiology.

Results: A total of 380 consecutive patients with a mean age of 1.9 ± 2.1 years (range, 1 month to 6 years) were included. Varied types of biventricular repair were customized individually. Follow-up was 90.4% complete, and the mean follow-up time was 3.4 ± 3.9 years. There were 17 (4.5%) early deaths and 7 (2.1%) late deaths. Preoperative pulmonary hypertension was the only risk factor for early mortality. Postoperative significant left ventricular outflow tract obstruction was present in 9 survivors. Patients with noncommitted ventricular septal defect had a longer crossclamp time, longer cardiopulmonary bypass time, and higher incidence of postdischarge left ventricular outflow tract obstruction. There were 4 reoperations, all of which were caused by subaortic left ventricular outflow tract obstruction. All of the pressure gradients were decreased to less than 20 mm Hg after the modified Konno procedure with an uneventful postoperative course.

Conclusions: Optimal results of varied types of biventricular repair for double outlet right ventricle have been acquired. Although noncommitted ventricular septal defect is technically difficult, the outcomes of patients are favorable. Late-onset left ventricular outflow tract obstruction is the main reason for reoperation but can be successfully relieved by the modified Konno procedure. (J Thorac Cardiovasc Surg 2014;148:817-24)

Double outlet right ventricle (DORV) is a congenital anomaly in which both the aorta and the pulmonary artery originate from the right ventricle, representing a broad spectrum of anatomic variants and associated malformations.[1,2] For many years, arguments have persisted on whether the lesion was better defined on the basis of (1) the connections between the arterial trunks and their supporting ventricle or (2) the presence of infundibular musculature supporting exclusively the leaflets of both arterial valves. To avoid misunderstanding, the 50% override rule was concisely used to define and diagnose DORV in the current study.

The pathophysiology of DORV varies from severe cyanosis to significant volume overload similar to a large ventricular septal defect (VSD) shunt. The optimal surgical approach is tailored to both the anatomic features and their physiologic consequences. A promising approach is highly varied and needs to be customized for individuals.[3]

Accompanying techniques have improved in the last decade, and a growing proportion of patients are undergoing biventricular repair.[4] However, the outcomes of anatomic repair for patients with DORV and noncommitted VSD are not as apparent. In addition, late postoperative subaortic obstruction has attracted attention. In the current study, we report our results among 380 patients with DORV presenting for biventricular repair at Fuwai Hospital (Beijing, PR China) and the surgical strategy for late-onset left ventricular outflow tract obstruction (LVOTO).

METHODS

Patients

Patients with DORV who underwent biventricular repair at Fuwai Hospital from January 2005 to December 2012 were included in this retrospective study. Diagnosis of DORV was made if both great arteries originated predominantly from the right ventricle with application of the "50% rule."[5] Patients were excluded if DORV was combined with

From the Pediatric Cardiac Surgery Center, State Key Laboratory of Cardiovascular Disease, National Center for Cardiovascular Disease and Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, People's Republic of China.

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Address for reprints: Shoujun Li, MD, Department of Cardiac Surgery, Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, 167 Beilishi Rd, Xicheng District, Beijing 100037, People's Republic of China (E-mail: drlishoujun@yahoo.com).

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Abbreviations and Acronyms

ANOVA = analysis of variance
DORV = double outlet right ventricle
IVR = intraventricular tunnel repair
LVOTO = left ventricular outflow tract obstruction
RVOT = right ventricular outflow tract
VSD = ventricular septal defect

atrioventricular septal defect, heterotaxy syndrome, atrioventricular discordance, or univentricular physiology. According to the location of VSD, patients were divided into group 1 (with noncommitted VSD), group 2 (with subaortic VSD), group 3 (with sub-pulmonic VSD), and group 4 (with doubly committed VSD). This study was approved by the ethics committee at Fuwai Hospital. The hospital gave us approval to waive the need for patient consent for publishing follow-up data on these patients.

Clinical Protocol and Surgical Technique of Biventricular Repair for Double Outlet Right Ventricle

Indication for varied approaches of anatomic repair depended on anatomic features, achieving biventricular repair for these patients. Intraventricular tunnel repair (IVR) to the aorta was performed in patients with VSD-type DORV; IVR to the aorta + right ventricular outflow tract (RVOT) reconstruction was performed in patients with tetralogy-type DORV; IVR + arterial switch operation or double root translocation was performed in patients with transposition-type DORV; and the réparation à l'étage ventriculaire or Rastelli procedure was performed when the pulmonary valve stenosis that was considered could not be enlarged. For DORV and noncommitted VSD, the choice of appropriate procedures was complex. The protocol of Fuwai Hospital is shown in Figure 1, *A*.

Pulmonary artery banding and modified Blalock–Taussig shunt were used as previous palliative approaches to restrict pulmonary plethora and alleviate cyanosis, respectively. At biventricular repair, all patients underwent operation with standard cardiopulmonary bypass, bicaval cannulation, and moderate hypothermia with cold potassium cardioplegic arrest. Intracardiac baffle was reconstructed by Dacron patch. VSD enlargement and chordae reattachment were selectively performed, substantially reducing the potential for subaortic stenosis and improving the baffle geometry. Both reducing the pulmonary valve regurgitation and avoiding further external conduit replacement were principles when reconstructing the RVOT. The techniques of the arterial switch operation or double root translocation were similar in reports from Morgan Stanley Children's Hospital[6] and our center.[7,8]

Clinical Protocol and Surgical Technique for Late-Onset Left Ventricular Outflow Tract Obstruction

At Fuwai Hospital, significant late postoperative LVOTO was defined as a trans-stenosis systolic pressure gradient of 30 mm Hg or greater. Indication for reoperation relieving late-onset LVOTO was stenosis-associating symptoms combined with a trans-stenosis systolic pressure gradient of 50 mm Hg or greater. Stenosis-associating symptoms included angina, syncope, and dyspnea. When the trans-stenosis systolic pressure gradient was greater than 75 mm Hg, surgical intervention was aggressively indicated regardless of symptoms.

The modified Konno was established as the procedure of choice to relieve postdischarge subaortic LVOTO after biventricular repair for DORV. Cardiopulmonary bypass techniques included continuous flow with bicaval cannulation, blood prime, and moderate hypothermia. Myocardial protection was achieved with antegrade single-dose blood cardioplegia. Previous intracardiac baffle, partial ventricular septal, and subaortic conus were carefully resected after right ventriculotomy. The incised area was limited to the upper part of the trabecular septum, and thus injury to the conduction system and major septal coronary arteries could be avoided. The ventricular septal was enlarged, and the new intracardiac baffle was reconstructed.

Data Collection and Definition

Patient demographics and clinical data were obtained from our local database. Ventricle function, size, and valve stenosis/regurgitation were assessed by echocardiography. During follow-up, patients were contacted by telephone or direct interview in our outpatient clinic every 3 to 6 months. A single cardiologist reviewed all previous echocardiograms and performed independent measurements. Pulmonary arterial hypertension was defined as mean pulmonary artery pressure more than 25 mm Hg. The severity of the valvular regurgitation was graded according to guidelines published by the American Society of Echocardiography.[9] Valvular regurgitation was considered significant when documented as moderate or severe, and ventricular dysfunction was defined as an ejection fraction less than 50%. In-hospital mortality was defined as both 30-day mortality and death any time after operation but before discharge. Postdischarge mortality was defined as death after 30 days or after discharge if the length of hospital stay was more than 30 days. Reoperation included only reoperations on the heart and excluded secondary closure of the sternum and revision for bleeding or mediastinitis.

Statistical Analysis

Continuous variables were presented as mean ± standard deviation or median with minimum and maximum range, and categoric variables were presented as percentage. Comparisons of variables were made using the Student *t* test, analysis of variance (ANOVA), chi-square test, or Fisher exact test. Bonferroni test was used in pairwise comparison. Time to death and time to postoperative LVOTO are displayed by Kaplan–Meier curves. Logistic regression (forward) was performed as multivariate analysis to investigate risk factors for early death, late death, and postoperative LVOTO. All entered variables were selected on the basis of clinical experience, univariate analysis, and previously published data. To find risk factors for early and late deaths, variables that entered the model included age less than 1 year at repair, preoperative pulmonary arterial hypertension, noncommitted VSD, aortic arch obstruction, previous palliation, pulmonary stenosis, and great arteries relationship. To find risk factors for postdischarge LVOTO, variables entered into the model included age less than 1 year at repair, noncommitted VSD, subaortic conus, pulmonary stenosis, and great arteries relationship. The level of significance was set at an alpha level of 0.05 or less. Analysis was conducted using SPSS version 17.0 (IBM-SPSS Inc, Armonk, NY) for Windows.

RESULTS

Patients' Characteristics and Anatomic Features

From January 2005 to December 2012, 380 consecutive patients with a mean age of 1.9 ± 2.1 years (range, 1 month to 6 years) underwent biventricular repair of DORV at Fuwai Hospital. The mean weight at biventricular repair was 12.2 ± 5.9 kg (range, 5.5-16 kg).

In our local database (not limited to 380 biventricular repairs), 25 palliative procedures were required in 25 patients (pulmonary artery banding and modified Blalock–Taussig shunt). There was no mortality in these patients, and 20 of them had undergone biventricular repair, who were included in this analysis. A total of 20 of 380 patients (5.3%) had a palliative procedure preceding complete repair. All of these patients required only 1 palliative